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EDITORIAL COMMENT

Infant Congenital Aortic Valve Stenosis

The Pendulum Swings*

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Thirty years have passed since the first report of successful balloon dilation of the aortic valve in an 8-year-old child with congenital aortic valve stenosis (1). Since that time balloon valvuloplasty has gradually gained favor over surgical aortic valvotomy as the procedure of choice in infants with congenital aortic valve stenosis (2). Most reports have centered on the outcomes of either balloon valvuloplasty or aortic valvotomy; there have been only a few studies that compare the outcomes of balloon valvuloplasty directly with aortic valvotomy (3). In this issue of the *Journal*, Siddiqui et al. (4) in their study from the Royal Children's Hospital, Melbourne is unique in that both therapies were offered concomitantly for a 14-year period giving the authors a unique perspective to compare the outcomes (4).

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Their "final answer" is that surgical valvotomy achieves better results than balloon valvuloplasty. They compared 37 patients undergoing balloon valvuloplasty in the neonatal and infant period to 86 patients undergoing surgical aortic valvotomy. The median age at the time of intervention was 27 days. The primary finding was that freedom from reintervention at 5 years was 27% after balloon valvuloplasty versus 65% after aortic valvotomy.

A very important point that I believe the authors from the Royal Children's Hospital, Melbourne, have emphasized is that surgical valvotomy is not simply incising the area of fused commissures with a knife. There are additional procedures that a surgeon may perform that may in fact enable a better outcome. These include resection of nodular dysplasia, thinning of the leaflets, re-creation of interleaflet triangles, and even the creation of neocommissures. Other surgeons have described excising myxomatous fibrous nodules and subvalvular resection if necessary. Patch enlargement of the noncoronary aortic sinus may be performed to give the thickened aortic leaflets more room to separate during systole. These additional procedures may lead to improved outcomes over balloon dilation in properly selected cases.

This paper adds to another recent publication, which appears to be pushing the pendulum back toward surgical intervention for congenital aortic valve stenosis. Dr. Brown et al. (5) from Indiana University published a similar study in 2012 in a slightly older patient population. They demonstrated that gradient reduction, degree of aortic insufficiency, and the need for re-intervention were worse after balloon dilation than surgical valvotomy. Of interest, however, is the fact that this study (Indiana) excluded neonates. Their surgical approach to neonates at that time was a transapical blunt dilation of the aortic valve with serial Hegar dilators (Cooper Surgical, Inc., Trumbull, Connecticut). This of course would be a very similar approach physiologically to a transcatheter balloon dilation. The rationale for this was that, "the aortic valve in neonates is frequently dysplastic, making identification of the commissures with an open approach using bypass difficult and imprecise." Their comment here was that the results from the closed surgical approach (in neonates) were satisfactory and not substantially different from those patients undergoing balloon dilation. The better results with surgical valvotomy were limited to the study population of older children.

One must recognize the limitations of the Melbourne and Indiana experiences. They are retrospective studies with no defined method of stratifying patients to balloon dilation or aortic valve surgery other than the surgeon and cardiologist preference. This can lend bias to the stratification of patients. At our institution it would be much more likely for a patient with a unicuspid aortic valve or an aortic valve that is severely dysplastic to be referred for balloon dilation over a surgical intervention. We had a recent patient present to us as a critically ill newborn with a gradient of 40+ mm Hg across the aortic valve, severe ventricular dysfunction, and very high left atrial pressure. We were very pleased that this patient could undergo a neonatal balloon dilation resulting in a decrease in the gradient to <10 mm Hg, no aortic valve insufficiency, and a dramatic drop in left atrial pressure. That type of intervention is quite appealing and saves the child from undergoing a sternotomy and cardiopulmonary bypass in the neonatal period when the left ventricle is functioning quite poorly. I still remember the occasional critical aortic stenosis patient with severely depressed function who would suddenly fibrillate during the sternotomy.

Another limitation of the Melbourne experience relates to the fact that they appear to have essentially abandoned balloon dilation in 2006. The past 7 years have seen steady improvements in techniques of cardiac catheterization including improvement in imaging in the catheterization

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laboratory, smaller French catheters, and higher pressure balloons. These technical advances probably will continue to occur within the world of interventional cardiology. In contrast the surgical armamentarium regarding the repair of neonatal aortic valves may have reached a "ceiling." One should not underestimate the importance of continued experience and improvement in the equipment used for interventional procedures. In the Boston experience over time the mortality of balloon valvuloplasty decreased from 22% (1985 to 1993) to 4% (1994 to 2002) (2). The accumulation of experience with a new procedure generally leads to improved results.

The Congenital Heart Surgeons' Society has also compared balloon dilation in neonates with surgical valvotomy and reached a conclusion that differed from the Royal Melbourne experience (3). This was a multi-institutional study of 110 neonates; 28 surgical and 82 balloon dilation (note the pendulum swing in numbers to balloon dilation). Although the survival and freedom from reintervention were similar there was a greater likelihood of important regurgitation with balloon valvuloplasty versus residual stenosis with surgical valvotomy. Interestingly, if one looks at the statistical analysis in this manuscript (which was carefully done) the outcomes differ depending on the depth of the analysis. Freedom from aortic valve related intervention stratified only by the type of initial treatment but not adjusted for any other factors led to a slight advantage of balloon dilation over surgical valvotomy. However, if one adjusts for differences in group characteristics as reflected by a propensity score derived from multiple logistical regression, there was a slight advantage to surgical valvotomy over balloon dilation. This comprehensive study (which in my review of the literature is the largest reported cohort of neonates and infants with critical aortic valve stenosis) demonstrates the complexity of the statistical analysis needed to arrive at therapeutic recommendations and guidelines.

In an interesting series from Birmingham, England, the authors evaluated 54 patients undergoing surgical aortic valvotomy in infancy (6). This was an institutional policy from 1989 to 2000 (no balloon dilation!). This group emphasized the fact that preoperative evaluation of *valve morphology* may enable selection of a group of patients in whom the results of surgery are excellent. They found that long-term outcomes were significantly better in infants in whom surgery results in trileaflet rather than bileaflet anatomy. Similar findings regarding the importance of valve morphology were observed by Hraška et al. (7) in 34 neonates undergoing surgical valvotomy. These conclusions, that valve morphology has a very important impact on outcome, are key. Patients who have nodules that can be resected or thickened leaflets that would benefit from thinning would be patients who can be better approached by a surgical technique. The patient with a unicuspid or severely dysplastic valve may be best served by balloon valvuloplasty.

Where is the pendulum? It appears to be swinging back toward surgical valvotomy in selected cases. However, I feel the true key is a critical evaluation of the *morphology* of the aortic valve prior to intervention. The approach to these patients should continue the collaborative efforts of most congenital heart centers where surgeons and interventional cardiologists both evaluate patients on a case-by-case basis. The use of 3-dimensional echocardiography can now identify the morphology of aortic valves much more clearly. Careful discussion by these 2 services should be able to determine whether there is an advantage to an operation or to balloon dilation in the catheterization laboratory. The patients who have a trileaflet valve may be the group who could be approached surgically and have an excellent long-term outcome as emphasized by the Birmingham and German groups. The group from Germany reports an interesting interdisciplinary consensus-based approach (7). If left ventricular function is depressed, a "gentle" balloon dilation (balloon size = 0.7 annulus size) is used as an intermediate step to stabilize the patient prior to surgical valvotomy. If the left ventricular function is not severely depressed, surgical valvotomy is the method of choice unless valve morphology is more favorable for a balloon dilation.

I believe we should be careful not to decide a priori that one approach is better than the other (i.e., one approach fits all neonates and infants with critical aortic valve stenosis). The paper from the Royal Children's Hospital of Melbourne and the recent paper from the Indiana University School of Medicine group both emphasize the fact that in selected cases improved results can be achieved with a surgical valvotomy. However, there are clearly many patients who may be better served by initial balloon dilation. We must not forget that both surgical valvotomy and balloon dilation are truly only palliative in nature and that probably the majority of these patients will require another reintervention. There is some advantage to avoiding a sternotomy if the patient is going to require multiple re-operations perhaps eventually culminating in aortic valve replacement and/or a Ross procedure. The pendulum continues to swing, but may now be stabilizing. As we improve our knowledge of how to care for these patients collaboration between the interventional cardiologist and the surgeon continues to be of paramount importance.

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